American Journal of Hematology 54:313 (1997)

Images in Hematology

Hemoglobin SC Disease

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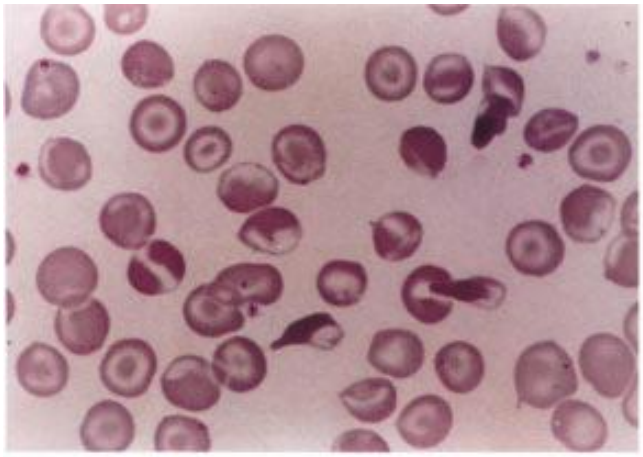
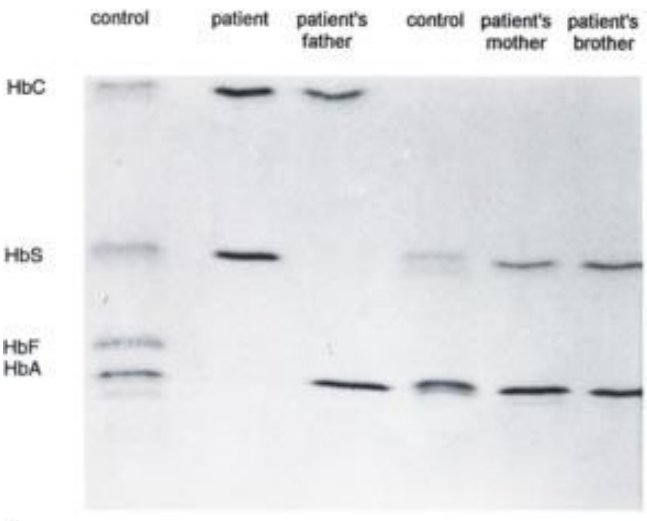


A

C



B

D

A 21-year-old black woman with no significant medical history presented with 3 days of jaundice, shortness of breath, and a hemoglobin of 1.5 g/dL. Physical exam revealed a suggestion of splenomegaly, a finding confirmed on abdominal ultrasound.

The peripheral blood film (image A) shows the characteristic features of hemoglobin SC disease. Most of the erythrocytes are ''target cells.'' Condensed hemoglobin aggregates (''hemoglobin crystals'') are also seen within the red cells, many of which are shrunken and distorted into pyrimidal and elongated forms.

Hemoglobin separation using isoelectric focusing (image B) shows both hemoglobin S (HbS) and hemoglobin C (HbC) to be present in the blood of the patient. The father is heterozygous for HbC and the mother is heterozygous for HbS.

An abdominal MRI scan (image C) reveals marked splenomegaly. There are several areas of decreased signal intensity characteristic of splenic infarction.

The presentation and findings are typical of hemoglobin SC disease complicated by acute splenic sequestration crisis. The patient was treated with packed red cell transfusion. Once she had recovered from the acute episode, splenectomy was performed to prevent future recurrences. At the time surgery, the spleen (image D) was noted to be moderately enlarged, weighing 470 g.

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